

## Two-chambered right ventricle: simulating two-chambered left ventricle

ALBRECHT BEITZKE, ROBERT H. ANDERSON<sup>1</sup>, JAMES L. WILKINSON, AND ELLIOT A. SHINEBOURNE

*From the Department of Cardiology, Children's Hospital, University of Graz, Austria; Department of Paediatrics, Cardiothoracic Institute, Brompton Hospital, London; and Department of Paediatrics, Royal Liverpool Children's Hospital, Liverpool*

**SUMMARY** Two cases are described of a most unusual variant of two-chambered right ventricle. In both the ventricular septal defect was between the distal chamber of the right ventricle and the left ventricle. However the extensive dividing 'septum' between proximal and distal parts of the right ventricle converted the latter, haemodynamically, into part of the left ventricle. In the first case the distal chamber supported the aorta in the left anterior position, the pulmonary artery arising from the proximal part of the right ventricle. In the second the pulmonary artery arose from the distal chamber and the aorta from the proximal chamber. Though in both the ventriculoarterial connection was double outlet right ventricle, functionally there was arterial concordance in case 1 and discordance in case 2. A further disconcerting feature was the resemblance of the distal right ventricular chamber to the rudimentary chamber of a univentricular heart of left ventricular type.

It is well known that a morphologically right ventricle can become divided by muscular bands into two discrete chambers. The distal part of the right ventricle may then resemble the rudimentary chamber seen in a univentricular heart of left ventricular type (Anderson *et al.*, 1976). Though usually associated with concordant ventriculoarterial connections, such a two-chambered right ventricle might exist with double outlet right ventricle. The distal subarterial part of such a two-chambered right ventricle could then communicate via a ventricular septal defect with the left ventricle. If the right ventricular 'septum' were sufficiently well developed, its distal chamber could haemodynamically and functionally become part of the left ventricle. Though we are unaware of previous descriptions of such malformations, we have now encountered two examples, which both give the appearance of 'two-chambered left ventricles'.

### Case 1

A male infant (birthweight 3.6 kg) was referred at the age of 3 months to another hospital, where a

diagnosis of ventricular septal defect was made. Despite treatment he did not gain weight and had several severe upper respiratory tract infections. At 5 months of age he was admitted to the Children's Hospital, University of Graz.

On examination his weight was below the third centile and he was acyanotic. Pulses were normal and systolic blood pressure was 100 mmHg. The liver was palpable 3 cm below the right costal margin. The second component of the second heart sound was accentuated and there was a grade 4/6 pansystolic murmur at the third left intercostal space. Chest x-ray film showed situs solitus with laevocardia; there was cardiomegaly with straight upper left cardiac border, and pulmonary plethora. The electrocardiogram showed sinus rhythm, a mean frontal QRS axis of +110°, right ventricular hypertrophy, and a shortened QT interval, the latter being attributed to digoxin therapy. Q waves were not seen in the precordial leads.

At cardiac catheterisation, two ventricles and both great arteries were entered, the ascending aorta via a ventricular septal defect, and the left ventricle via right atrium, patent foramen ovale, and left atrium. The right ventricular (60/3 mmHg) and pulmonary artery pressures (45/15 mmHg) were raised but were below systemic level. The systolic pressure in the ascending aorta was the same as in

<sup>1</sup>Robert H. Anderson is supported by the Joseph Levy Foundation together with the British Heart Foundation.

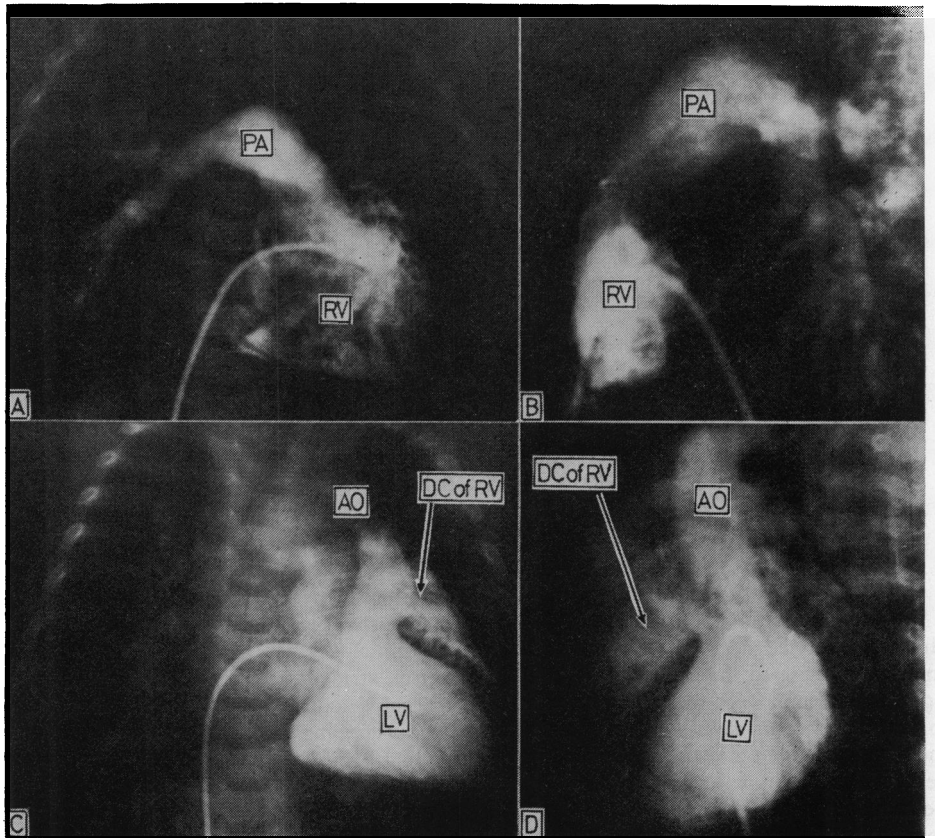


Fig. 1 Case 1. Angiocardigrams. Fig. 1A shows an injection into the morphologically right ventricle in the frontal projection; Fig. 1B is the same injection in the lateral projection. The right ventricle (RV) gives rise to the pulmonary artery (PA). Fig. 1C and 1D show a left ventricular injection in frontal (C) and lateral (D) projections. The left ventricle (LV) is in continuity with the distal chamber of a two-chambered right ventricle (DC of RV), and the aorta (AO) overrides the septum between the chambers.

the left ventricle (94 mmHg). Oximetry showed a large left-to-right shunt and a small right-to-left shunt at ventricular level. The  $\dot{Q}_p/\dot{Q}_s$  ratio was 2.4:1.

Subsequent catheterisation 12 months later, performed because of recurrent chest infections and persistent cardiomegaly, showed almost normal right ventricular (35/5 mmHg) and pulmonary artery (35/12 mmHg) pressures but the left-to-right shunt had decreased, with a  $\dot{Q}_p/\dot{Q}_s$  ratio of 1.7:1. At this time there were no signs of cardiac failure and the chest infections were found to be related to hypogammaglobulinaemia.

The angiocardigraphic features from the two studies have been combined for ease of presentation.

The presence of situs solitus was confirmed, the catheter entering a morphologically right atrium

through a right-sided inferior vena cava. A right-sided anterior ventricle was entered from the right atrium and this chamber was triangular and coarsely trabeculated (Fig. 1A). The pulmonary artery arose from this chamber and there was infundibular tissue between the tricuspid and pulmonary valves (Fig. 1B). A left-sided and posterior ventricular chamber was entered via the left atrium and left-sided atrioventricular valve. This chamber was smooth-walled and ovoid (Fig. 1C and 1D). The injection into the left-sided ventricular chamber also disclosed the presence of a third discrete chamber within the ventricular mass (Fig. 1C). This chamber gave rise to a left-sided anterior aorta, with aortic valve at the same level as the pulmonary valve and separated by infundibular tissue from both atrioventricular valves (Fig. 1C). A selective

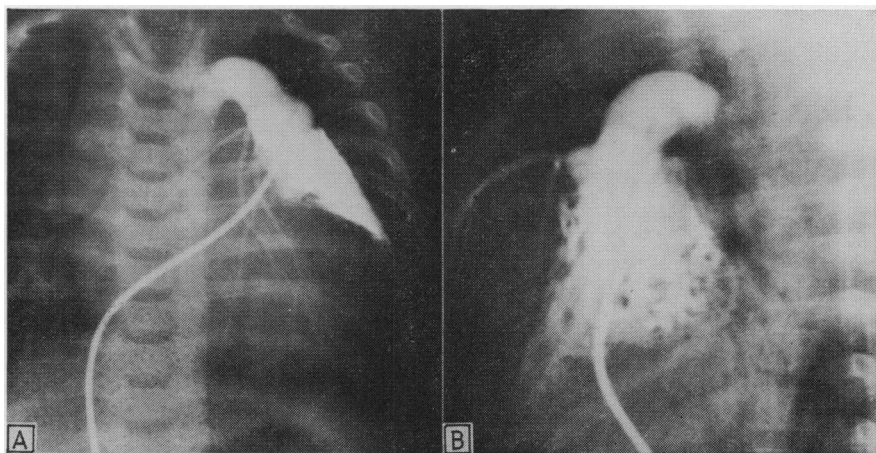


Fig. 2 Case 1. Angiocardiogram with selective injection into the distal chamber of the right ventricle, viewed in the frontal (Fig. 2A) and lateral (Fig. 2B) projections. The chamber has a trabecular zone with right ventricular characteristics and supports part of the aorta.

injection into this chamber (Fig. 2A and 2B) showed that it possessed a coarse trabecular zone and did indeed give rise to the aorta.

The angiocardiograms were interpreted as showing the presence of situs solitus, atrioventricular concordance, with double outlet from the right ventricle. The third chamber was interpreted as being part of the morphologically right ventricle that had become sequestered in such a way as to communicate mainly with the left ventricle. There was a sub-aortic ventricular septal defect between the two ventricles and the aorta was left-sided. There was a bilateral infundibulum.

## Case 2

This baby girl was admitted to the Royal Liverpool Children's Hospital at the age of 8 days with a history of cyanosis and dyspnoea from the second day of life. On examination she was found to be in cardiac failure with moderate cyanosis. There were no cardiac murmurs. The chest x-ray film showed cardiomegaly, a narrow pedicle, and pulmonary plethora. The electrocardiogram showed sinus rhythm with left ventricular preponderance abnormal for age. Angiocardiograms were interpreted as showing complete transposition with ventricular septal defect and overriding of the pulmonary artery. A balloon atrial septostomy was performed, after which progress was initially satisfactory. However the infant then deteriorated rapidly and died at the age of 2½ weeks.

At necropsy the aorta and pulmonary artery were side by side, with pulmonary artery much larger

than aorta (Fig. 3A). There was isthmal hypoplasia. An initial segmental diagnosis was made of situs solitus, atrioventricular concordance, and ventriculo-arterial discordance (complete transposition), with the aorta rising from the morphologically right ventricle (Fig. 3B) and the pulmonary artery from the left ventricle (Fig 3D) overriding the ventricular septal defect. Further examination, however, showed that though an interventricular septum was present to the right of the pulmonary artery, the anterior descending branch of the left coronary artery was to the left of the pulmonary artery running in the interventricular groove. Dissection in the ventricular wall beneath the pulmonary artery then disclosed the presence of a third chamber in the ventricular mass (Fig. 3C). This chamber had the trabecular characteristics of the right ventricle, was separated by the infundibular septum and apical trabeculae from the rest of the right ventricle, and from the left ventricle by the anterior trabecular septum reinforced by the trabecula septomarginalis. This subpulmonary chamber was in free communication with the left ventricle via the subpulmonary ventricular septal defect. In contrast, it was almost completely separated from the proximal part of the right ventricle, only a probe patent passage existing between the two parts of the morphologically right ventricle. The pulmonary valve was separated from the mitral valve by the ventriculo-infundibular fold, which was partially attenuated to permit minimal pulmonary mitral fibrous continuity.

The final segmental diagnosis was situs solitus, atrioventricular concordance, and double outlet right ventricle with subpulmonary ventricular septal

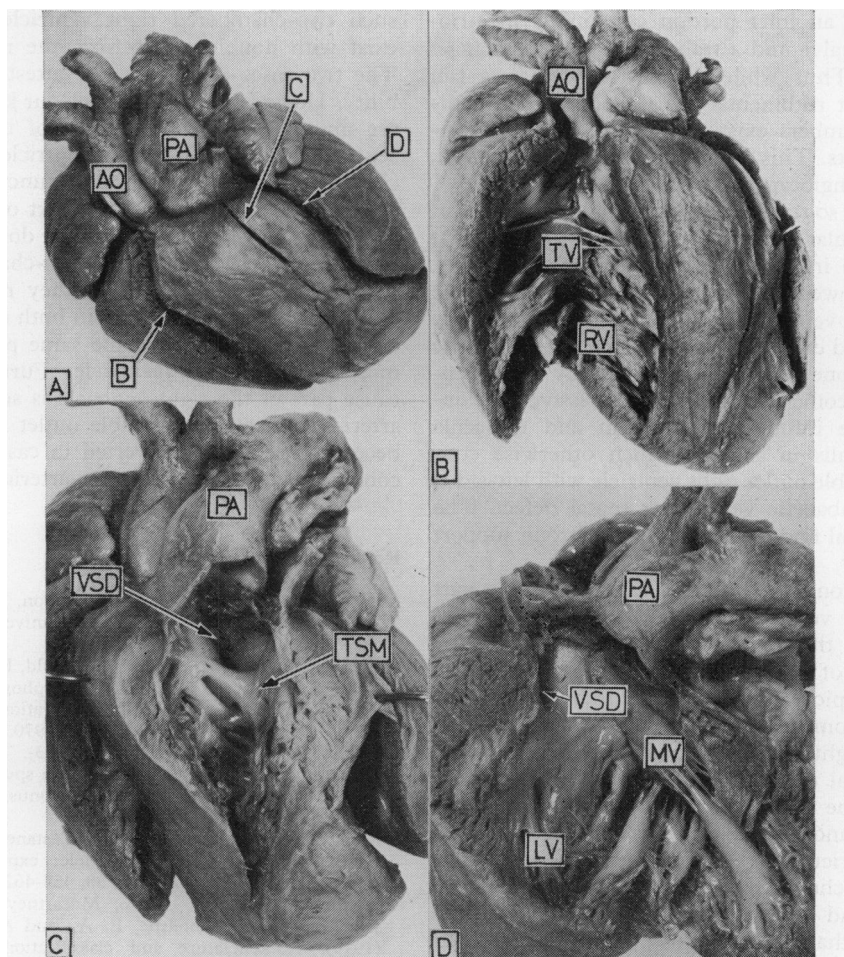


Fig. 3 Case 2. Photographs of the necropsy specimen. Fig. 3A shows the anterior view of the heart, with the aorta (AO) and pulmonary artery side-by-side. Fig. 3B, 3C, and 3D show the chambers of the heart, viewed through cuts B, C, and D in Fig. 3A. Fig. 3B shows the proximal chamber of the right ventricle (RV), which receives the tricuspid valve (TV) and gives rise to the aorta (AO). Fig. 3C shows the distal chamber of the right ventricle, separated by an almost complete muscular partition from the proximal chamber but in free communication with the left ventricle through the subpulmonary ventricular septal defect (VSD). Note the trabecula septomarginalis (TSM) which forms the lower rim of the defect. Fig. 3D shows the left ventricle (LV), which receives the mitral valve (MV) and supports part of the pulmonary artery (PA), which overrides the defect in the septum (VSD) between the left ventricle and the distal chamber of the right ventricle.

defect, the important additional anomaly being a two-chambered right ventricle with the distal chamber in free communication with the left ventricle.

### Discussion

These cases are of considerable interest in that they

possess two ventricles, but also an additional chamber within the ventricular mass, possessing both infundibular and trabecular portions. In this respect, the third chamber is analogous with the rudimentary chamber often seen in univentricular heart of left ventricular type. However, in these hearts there is no question that two ventricles are present,

each having an inlet portion containing an atrio-ventricular valve and a trabecular zone (Tynan *et al.*, 1979). Thus while univentricular hearts can exist without rudimentary chambers, so can rudimentary chambers exist in the presence of biventricular hearts. This is an important consideration, since the angiocardigraphic features of case 1 (Fig. 2) were so reminiscent of the outlet chamber of a univentricular heart of left ventricular type that this was the initial diagnosis. Segmental analysis, however, showed that this was not the case, discrete right and left ventricles being unequivocally present. Our proposed explanation for this is that part of the trabecular zone of the morphologically right ventricle had become sequestered by excessive development of the infundibular septum and trabecula septomarginalis in a heart which otherwise consisted of double outlet right ventricle with left-sided aorta and subaortic ventricular septal defect. The morphological findings in case 2 add strong support to this theory.

In this second case, there is little doubt that part of the right ventricle has effectively been incorporated into the morphologically left ventricle by overgrowth of the infundibular septum and its fusion with apical right ventricular trabeculae. Thus, this case is comparable to a double outlet right ventricle with right-sided aorta and subpulmonary ventricular septal defect (Taussig-Bing malformation) in which the right ventricle has become two-chambered and the distal chamber communicates via the ventricular septal defect with the left ventricle. Two-chambered right ventricles are more usually found with concordant ventriculoarterial connections than with double outlet ventricles. Such two-chambered right ventricles, however, are frequent in Fallot's tetralogy (Rowland *et al.*, 1975), and, since there is a spectrum of malformed hearts between the tetralogy of Fallot and double outlet right ventricle (Goor and Edwards, 1973; Anderson *et al.*, 1974), it is not unreasonable to expect that

such two-chambered right ventricles should also exist with double outlet from the right ventricle. The two cases described here attest to this possibility. However, to the best of our knowledge they are unique, since the presence of the ventricular septal defect between the left ventricle and the distal chamber of the right ventricle functionally incorporated the distal chamber as part of the *left* ventricle. Thus, though there is no doubt that morphologically the hearts are two-chambered *right* ventricles, haemodynamically they represent two-chambered left ventricles, as in both cases the rudimentary chamber was at the same pressure as the morphologically left ventricle. Furthermore, because part of the right ventricle is sequestered the arterial connection of double outlet right ventricle becomes effectively converted in case 1 to arterial concordance and in case 2 to arterial discordance.

## References

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Requests for reprints to Dr R. H. Anderson, Department of Paediatrics, Cardiothoracic Institute, Brompton Hospital, Fulham Road, London SW3 6HP.